Hyperparathyroidism-jaw tumour syndrome detected by aggressive generalized osteitis fibrosa cystica

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Summary

Severe hyperparathyroidism can affect bone metabolism and be in the origine of multiple brown tumours (generalized osteitis fibrosa cystica). When associated with fibro-ossifying tumours of the jaw, it realizes a rare genetic syndrome referred as Hyperparathyroidism-jaw tumour HPT-JT.
We report the case of a patient we treated for HPT-JT, and literature review.

KEY WORDS: hyperparathyroidism; osteitis fibrosa cystica; ossifying fibroma.

Introduction

Brown tumours, also called osteitis fibrosa cystica, are relatively rare non-neoplastic osteolytic lesions of bones that appear in an advanced stage of hyperparathyroidism and involve mostly axial skeleton and femora. These lesions may produce awful pain and pathologic fractures (1-3).
Association of primary hyperparathyroidism and ossifying fibroma of the jaw is seen in a rare hereditary syndrome referred as hyperparathyroidism-jaw tumour HPT-JT.
We report a case of a female patient who was treated for generalized osteitis fibrosa cystica related to a primary hyperparathyroidism associated to an ossifying fibroma of the jaw.

Case report

A 41-year-old woman suffering from diabetes for 8 years under glibenclamide, treated for pulmonary tuberculosis 3 years ago, who underwent surgery for radial bone osteitis 12 years ago, toothless, and without history of radiotherapy or family disease, was referred to our department for a palpable but indolent tumour of the left maxilla (Figure 1). Orthopantomography and Computed tomography (Figure 2) found a well-defined osteocondensing lesion in the left maxillary tuberosity and a silent premaxillar cyst. We removed the maxillary tuberosity tumour which revealed an ossifying fibroma and we decided to follow up the premaxillar cyst. Six months later, the patient returned to the hospital suffering from an extreme fatigue, a severe weight loss (24 kg in 6 months), and also intolerable spontaneous pain of bones and joints involving especially right arm and leg. Clinical examination found a frontal bone tumour, dorsal and lumbar spinous processes pains and a cervical node attached to the right lobe of the thyroid. Bones radiography revealed generalized demineralization, multiple osteolytic lesions of the skeleton affecting long bones and frontal bone (Figure 3)

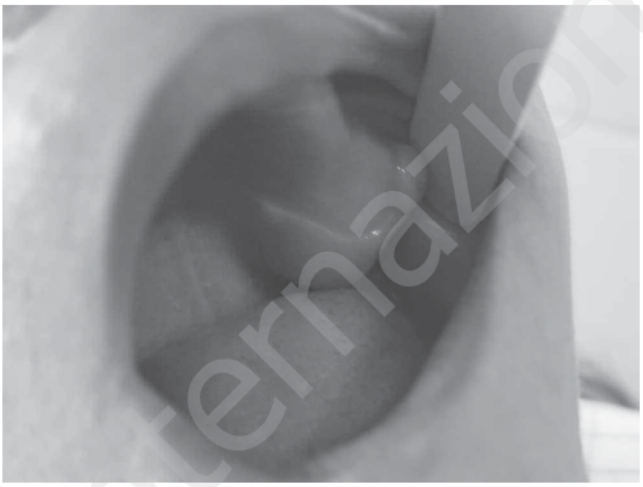

Figure 1 - The tumour of the left maxilla for which the patient was first seen.

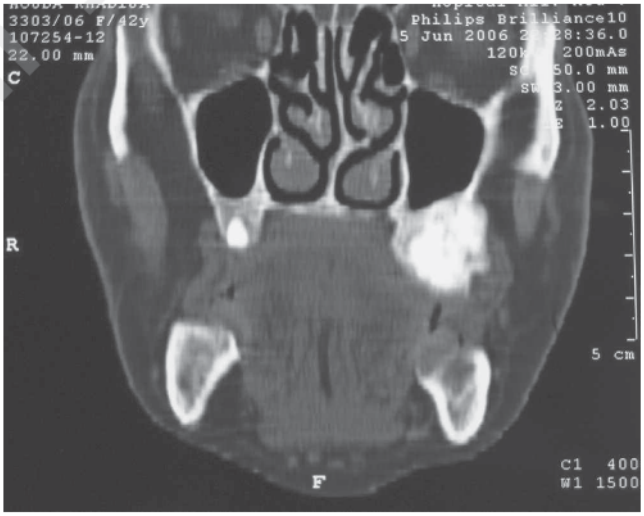

Figure 2 - The CT is showing an osteocondensing lesion of the left maxillary tuberosity.